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ORIGINAL ARTICLE

Differentiating presacral masses in anorectal malformations and isolated sacrococcygeal teratomas

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Abstract

Purpose Presacral masses associated with anorectal malformations (ARM) are most frequently dermoid or teratomas. Sacrococcygeal teratoma (SCT), in isolation, is a different condition. There are limited data comparing the two. The aim of this study was to compare presacral masses associated with ARM and isolated SCTs.

Methods A retrospective review was performed from 1979 to 2018. Patients with presacral masses and associated ARM ($n=39$) were identified and compared to patients with isolated SCTs ($n=32$).

Results Those with presacral mass and ARM had a lower proportion of immature and mixed teratomas by pathology and a longer time to mass resection. They had higher percentages of sacral anomalies (76% versus 9%), tethered cord (50% versus 6%), and other anomalies (51% versus 9%). For long-term bowel function, patients with presacral mass and ARM were more likely to be fecally incontinent and require enemas (59% versus 11%). Regarding isolated SCTs, 4% recurred with a time to recurrence of 7 months.

Conclusions Compared to patients with isolated SCT, patients with presacral mass and ARM had lower grade tumors. In addition, they had a higher proportion of sacral defects and other congenital defects, as well as worse bowel function outcomes.

Keywords Presacral mass · Anorectal malformation · Sacrococcygeal teratoma · Bowel function outcomes

Introduction

Presacral masses occur in about 1 in 40,000 births [1] and demonstrate a wide spectrum of presentation and associated anomalies. The majority of presacral masses are dermoids, teratomas, lipomas, anterior meningoceles, or a combination thereof. They are frequently associated with a sacral defect and anorectal malformation (Currarino triad) [2]. The overwhelming majority are benign by histology.

Isolated sacrococcygeal teratomas that originate in the coccyx, are stratified according to the Altman classification [3], and have a significant malignant potential. These tumors are usually identified prenatally or at birth and are resected early in life [3].

There are limited data [4] comparing presacral masses associated with ARM and isolated sacrococcygeal teratomas. The aim of this study was to compare these two entities and their respective clinical outcomes.

Methods

Study design

A retrospective chart review was performed at a tertiary care children's hospital. Patients with presacral masses associated with anorectal malformations were obtained from the author's colorectal database spanning 1979–2018. Patients with isolated sacrococcygeal teratomas were identified from the radiology database spanning 2006–2019. The study

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was approved by the Colorado Institutional Review Board (COMIRB # 17-1670).

Data review

Patient characteristics included gender, age at surgery, type of anorectal malformation, sacrococcygeal teratoma classification, associated diagnoses (sacral defect, spinal abnormality, urogenital abnormality, musculoskeletal anomaly, cardiac anomaly, gastrointestinal anomaly), mass size, and family history of presacral mass. Sacrococcygeal teratoma classification was based on the Altman classification [3] where type I is entirely outside the body, type II is mostly outside the body, type III is mostly inside the body, and type IV is entirely inside the body.

Operative details included pathology, time to surgery, operative procedure, completeness of resection, tumor spillage, and whether coccygectomy was performed. Outcomes examined included recurrence (including whether patient received chemotherapy) and bowel function outcomes. Bowel function outcomes were excluded for those less than 3 years of age as this was prior to toilet training.

Statistical analysis

Patients with presacral mass and anorectal malformation (ARM) ($n=39$) were compared to patients with isolated sacrococcygeal teratomas (SCT) ($n=32$). Median with interquartile range was used for continuous variables, and proportions were used for categorical variables. Significance testing was performed using Wilcoxon rank-sum test and Fisher's exact test as appropriate. Logistic regression was performed with continence outcomes (requiring enemas or not) as the dependent variable. Independent variables examined included gender, type of ARM, type of SCT, sacral abnormality, tethered cord, and mass size. However, there was not enough information regarding mass size in the ARM group to perform regression. All analysis were performed using Stata.

Results

Patient characteristics

The majority of patients in both groups were female (67% in ARM versus 61% in SCT, $p=0.8$). Although not statistically significant, there was a higher proportion of patients with a family history of a presacral mass in the ARM group (4/36, 11%) compared to the SCT group (1/31, 3%) ($p=0.4$) (Table 1).

Types of anorectal malformations included rectal stenosis (11/38, 29%), imperforate anus (5/38, 13%), rectoperineal fistula (4/38, 11%), rectovestibular/rectourethral fistula (10/38, 26%), cloaca (6/38, 16%), and cloacal exstrophy (2/38, 5%).

Table 1 Patient characteristics comparing those with presacral masses associated with anorectal malformations (ARM) and sacrococcygeal teratomas (SCT)

	ARM with presacral mass ($n=39$)	SCT ($n=32$)	p value
%Female	67% (26/39)	61% (17/28)	0.8
Family history of presacral mass	11% (4/36)	3% (1/31)	0.4
Other anomalies[a]	51% (20/39)	19% (6/31)	**0.01**
Tethered cord	50% (19/38)	6% (2/32)	**< 0.001**
Sacral abnormality			**< 0.0001**
Hemisacrum	76% (22/29)	9% (3/32)	
Bifid sacrum	48% (14/29)	3% (1/32)	
Agenesis	10% (3/29)	0% (0/32)	
Other	0% (0/29)	3% (1/32)	
	14% (4/29)	3% (1/32)	

Significance is a p value less than 0.05

[a]Other anomalies: renal agenesis, dysplastic kidney, neurogenic bladder, vesicoureteral reflux, esophageal atresia, omphalocele, clubfoot, absent radius, lumbar kyphosis, uterus agenesis, hemivagina, undescended testicles, double outlet ventricle, atrial septal defect, dextrocardia

The majority of SCTs were type 2 (10/22, 45%) followed by type 3 (7/22, 32%), type 4 (3/22, 14%), and type 1 (2/22, 9%) (Fig. 1).

There was a significantly higher proportion of sacral abnormalities (76% versus 9%, $p<0.0001$), tethered cord (50% versus 6%, $p<0.001$), and associated anomalies (51% versus 19%, $p=0.01$) in the ARM group compared to the isolated SCT group. Of the sacral abnormalities in the ARM group, the majority had a hemisacrum (14/29, 48%) followed by an otherwise unspecified abnormal sacrum (4/29, 14%), and bifid sacrum (3/29, 10%) (Table 1). In the ARM group, the other anomalies included renal dysgenesis (3/39), vesicoureteral reflux (6/39), tracheoesophageal fistula (3/39), omphalocele (1/39), limb anomalies (3/39), hemivagina (6/39), and cardiac anomalies (4/39). In the isolated SCT group, the other anomalies included neurogenic bladder (4/31), clubfoot (2/31), dysplastic kidney (1/31), and ocular teratoma (1/31).

Operative characteristics

The mass size in the isolated SCT group (7.5 (IQR 3.5–11) cm) was significantly larger than the ARM group (5 (IQR 3–5) cm), $p=0.02$) (Table 2). To that end, in the isolated SCT group, 3/32 underwent partial resections at the time of delivery due to concern for hemorrhage. Those in the ARM group had a significantly longer time to mass resection

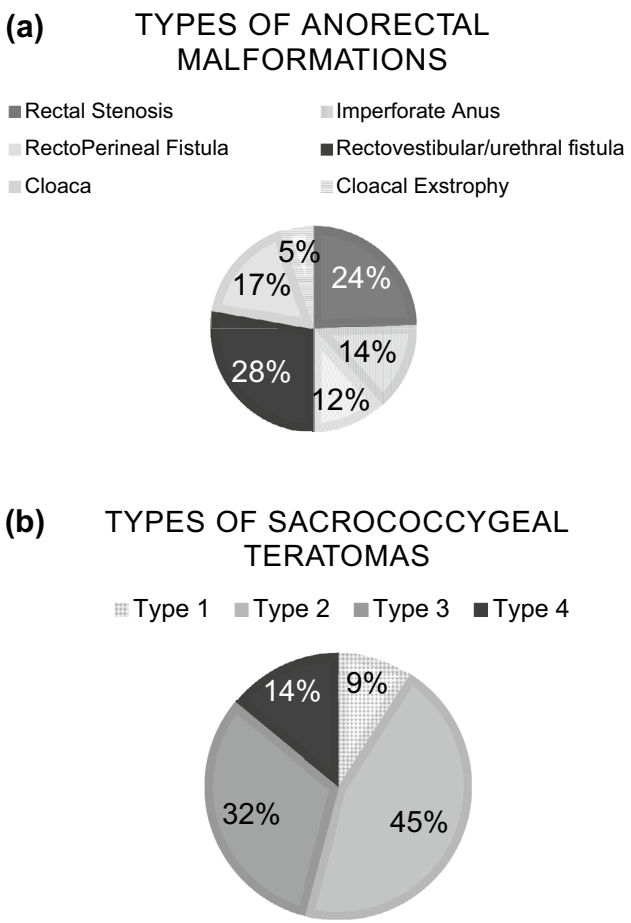

Fig. 1 **a** Types of anorectal malformations. **b** Types of sacrococcygeal teratomas

Table 2 Operative characteristics comparing those with presacral masses associated with anorectal malformations (ARM) and sacrococcygeal teratomas (SCT)

	ARM with presacral mass	SCT	*p* value
Median age at mass resection (days)	677 (181–1402)	8 (IQR 2–284)	< 0.0001
Mass size (cm)	5 (3–5)	7.5 (3.5–11)	0.02
Histology	37% (10/27)	0%	
Lipomeningocele	59% (16/27)	46% (11/24)	
Mature teratoma	0%	25% (6/24)	
Immature teratoma	0%	29% (7/24)	
Mixed teratoma			

with a median age at resection of 677 (IQR 181–1402) days compared to 8 (IQR 2–284) days ($p < 0.0001$). In the ARM group, 37/39 underwent mass resection at the time of posterior sagittal anorectoplasty (PSARP).

Operative details regarding tumor spillage was not available for the ARM patients and performance of coccygectomy is not part of the surgical technique, as the coccyx is not present in the majority of the patients (22/29). In the isolated SCT group, tumor spillage occurred in 22% (5/23) of cases and coccygectomy was performed the majority of the time (88%, 21/24).

The histology differed between the two groups. In the ARM group, the majority of masses were mature teratomas (59%, 16/27) and 37% (10/27) were lipomeningoceles. In the isolated SCT group, the majority were mature teratomas (46%, 11/24) followed by mixed teratoma (29%, 7/24), and immature teratoma (25%, 6/24) (Table 2).

Outcomes

In the isolated SCT group, 19% received neoadjuvant or adjuvant chemotherapy. At a median follow-up of 2.8 years, 1/24 (4%) had recurred at 7 months.

Those in the isolated SCT group had improved bowel function outcomes compared to the ARM group ($p = 0.003$). At a median of 2.7 years of follow-up, the majority of patients in the isolated SCT group (6/15, 40%) were stooling normally without medications and 47% (7/15) required laxatives. Only 13% (2/15) were fecally incontinent and required enemas. At a median of 5.5 years of follow-up in the ARM group, the majority (14/21) were fecally incontinent but were kept clean with the use of enemas. Otherwise 29% (6/21) required laxatives, and only 5% (1/22) did not require any medication.

On bivariate analysis, the ARM group had 22 times greater odds (95% CI 4, 117; $p < 0.0001$) of being fecally incontinent when compared to the isolated SCT group. Patients with a sacral abnormality had 17 times greater odds (95% CI 3, 103; $p = 0.002$) of suffering from fecal incontinence than those with a normal sacrum. Patients with a tethered cord had six times greater odds (95% CI 1.5, 27; $p = 0.01$) of being fecally incontinent than those without tethered cord. As the complexity of the ARM increased (from rectal stenosis to imperforate anus, to perineal fistula, to vestibular/urethral fistula, to cloaca), the odds of being incontinent increased by two times for every increase in complexity of ARM (95% CI 1, 4; $p = 0.03$). On multivariable analysis adjusting for type of presacral mass, sacral abnormality, tethered cord, and type of ARM, no variable was significant (Table 3).

Table 3 Unadjusted odds ratios for fecal incontinence

	OR (95%CI)	*p* value
ARM vs. SCT	22 (4, 117)	< 0.0001
Sacral abnormality	17 (3, 103)	0.002
Tethered cord	6 (1.5, 27)	0.01
Complexity of ARM	2 (1, 4)	0.03

ARM anorectal malformation, *SCT* sacrococcygeal teratoma

Discussion

Presacral masses associated with anorectal malformations and isolated sacrococcygeal teratomas represent different entities with different etiology, malignant potential, and associated defects. Presacral masses associated with ARM are generally resected later in life. Presacral masses associated with ARM have a higher percentage of associated anomalies specifically sacral abnormalities and tethered cord. Lastly, those with presacral masses associated with ARM have worse bowel function outcomes compared to those with isolated SCT.

In comparison to other studies, our study demonstrated a more varied distribution of anorectal malformations. We found that anorectal stenosis represented 24% of the ARMs which is lower than reported in other studies where the proportion of anorectal stenosis has ranged from 41 to 88% [1, 4, 5]. While anorectal stenosis should raise suspicion for presence of an underlying presacral mass, our study found that presacral masses can occur in a variety of other anorectal malformations. Regarding isolated SCTs, our study demonstrated a higher proportion of type 3 (45%) and a lower proportion of type 1 (9%) compared to other studies where type 3 ranged from 4% to 14% and type 1 ranged from 33 to 45% [3, 4, 6]. The reason for the higher proportion of type 3 SCTs in our population is unclear.

Anomalies associated with presacral masses and ARM are varied. Reports of tethered cord range from 27 to 70% [5, 6] compared to 50% in our data set. In patients with ARM, the proportion with a normal sacrum range from 12 to 17% [4, 6] compared to 24% in the current study. The classic description of Currarino triad includes a hemisacrum, which was present in 48% of our ARM group. However, it should be noted that 24% of patients had other sacral abnormalities, depending on the location of the mass. Masses located in the midline typically result in a bifid sacrum or hemisacrum (Fig. 2).

Our findings mirror the existing literature in that isolated SCTs are often resected at younger patient age [7] and are typically larger in size when compared to presacral masses associated with ARM [4]. As in our study, the majority of presacral masses associated with ARM and isolated SCTs are mature teratomas, with the proportions ranging from 67 to 81% in the ARM group and 59–82% in the SCT group [1, 3, 4, 7]. The other presacral masses in ARM patients consisted of lipomyelomeningoceles.

This is the first study to examine bowel function outcomes between the two groups. Not surprisingly, those with presacral masses associated with ARM had worse bowel outcomes, even when they originally had a good functional prognosis type of malformation (recto-perineal fistula). The type of anorectal malformation contributes to this as demonstrated by the increasing odds of requiring enemas as the types of malformation progress from anorectal stenosis to cloaca. Furthermore, sacral abnormalities were present in patients with presacral masses associated with ARMs in a much higher proportion compared to the isolated SCT group. Previous studies have demonstrated the importance of a normal sacrum to continence outcomes [8, 9].

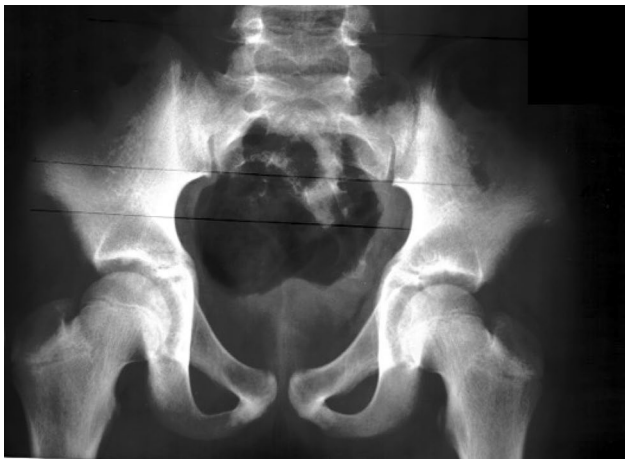

Fig. 2 Radiograph of hemisacrum

Limitations of this study include its retrospective nature including missing data and a small sample size. However, it represents one of the larger studies in the literature comparing these two disease entities and provides valuable insight into differences in presentation and outcome. Long-term prospective studies are still needed and they will better guide clinicians on follow-up required for patients with presacral masses associated with anorectal malformation. In conclusion, compared to patients with isolated SCT, patients with presacral mass and ARM had a higher proportion of sacral defects and other congenital defects. In addition, they had lower grade tumors and poorer bowel function outcomes.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Human and animal rights All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee (include name of committee + reference number) and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was not obtained as identifiable information was not included in the manuscript.

References

1. Bartels SA, van Koperen PJ, van der Steeg AF et al (2011) Presacral masses in children: presentation, aetiology and risk of malignancy. Colorectal Dis 13:930–934
2. Currarino G, Coln D, Votteler T (1981) Triad of anorectal, sacral, and presacral anomalies. AJR Am J Roentgenol 137:395–398
3. Altman RP, Randolph JG, Lilly JR (1974) Sacrococcygeal teratoma: American Academy of Pediatrics Surgical Section Survey-1973. J Pediatr Surg 9:389–398
4. Halleran DR, Vilanova-Sanchez A, Reck CA et al. (2018) Presacral masses and sacrococcygeal teratomas in patients with and without anorectal malformations: a single institution comparative study. J Pediatr Surg 54(7):1372–1378
5. Emans PJ, Kootstra G, Marcelis CL et al (2005) The Currarino triad: the variable expression. J Pediatr Surg 40:1238–1242
6. AbouZeid AA, Mohammad SA, Abolfotoh M et al (2017) The Currarino triad: what pediatric surgeons need to know. J Pediatr Surg 52:1260–1268
7. Dirix M, van Becelaere T, Berkenbosch L et al (2015) Malignant transformation in sacrococcygeal teratoma and in presacral teratoma associated with Currarino syndrome: a comparative study. J Pediatr Surg 50:462–464
8. Dewberry L, Pena A, Mirsky D et al (2018) Sacral agenesis and fecal incontinence: how to increase the index of suspicion. Pediatr Surg Int 35:239–242
9. Pena A (1995) Anorectal malformations. Semin Pediatr Surg 4:35–47